# PARENT FACT SHEET

## DISORDER

# Nonketotic Hyperglycinemia (NKH)

## CAUSE

Nonketotic Hyperglycinemia (NKH) is an inherited condition in which the body is unable to breakdown and process some of the building blocks of protein (amino acids). It is considered an amino acid condition because it can lead to high levels of the amino acidglycine in the body. There are several forms of NKH, which differ in regards to disease severity and age of onset. The symptoms and long term outcome of each form vary widely. In some forms of NKH, detecting the condition early and beginning proper treatment may help prevent or delay some of the severe health outcomes associated with the condition. It is important to remember that every child diagnosed with NKH is affected differently.

## IF NOT TREATED

Signs of NKH can begin any time from infancy to adulthood and include:

- Sleeping longer or more often
- Weak muscle tone
- Wandering eye movements
- Abnormal jerky movements
- Difficulty feeding
- Difficulty breathing
- Developmental delay
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The severity and onset of NKH differs depending on the form.

## TREATMENT OPTIONS

There is no cure for nonketotic hyperglycinemia (NKH). Sodium Benzoate and dextromethorphan are the prescribed treatments to help your baby's body get rid of excess glycine and reduce seizures and symptoms of severe nonketotic hyperglycinemia (NKH). However, your baby's doctor may recommend anti-epileptic drugs to reduce seizures also. Occupational, physical, and speech therapies are indicated to help your child reach their full potential.

## IF TREATED

Not all children with NKH will respond to treatment. Others may have great results.

Babies with NKH typically have varying degrees of developmental delay and intellectual disability.